

Nonmucinous Cystadenoma of the Pancreas with Pancreatobiliary Phenotype and Ovarian-like Stroma. Case Report of a Recently Described Entity

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Abstract The World Health Organization (WHO) classification of cystic pancreatic tumors defines mucinous cystic neoplasms as large, thick-walled, septated cysts with no communication with the main pancreatic duct, associated with the presence of ovarian-like stroma. Serous cystic neoplasms, on the contrary, present with a cuboidal or simple squamous epithelium and fibrous or hyalinized stroma without ovarian-like stroma. We present the case of a 23-year-old woman presenting with abdominal pain related to a new type of cystic pancreatic neoplasm characterized by non-mucinous pancreaticobiliary cystic epithelium and ovarian-like stroma that was surgically removed.

Keywords: *pancreas, pancreatic cysts, cystadenoma*

Cite This Article: Adriana Fabiola Romano-Munive M.D., Mario C Peláez M.D., Jorge Hernández-Calleros M.D., Carlos Chan-Núñez M.D., and Luis Uscanga-Domínguez M.D., “Nonmucinous Cystadenoma of the Pancreas with Pancreatobiliary Phenotype and Ovarian-like Stroma. Case Report of a Recently Described Entity.” *American Journal of Medical Case Reports*, vol. 3, no. 12 (2015): 391-393. doi: 10.12691/ajmcr-3-12-2.

1. Introduction

The World Health Organization (WHO) classification of cystic pancreatic tumors defines mucinous cystic neoplasms (MCNs) as large, thick-walled, septated cysts with no communication with the main pancreatic duct [1]. Histologically, these cysts are characterized by the presence of ovarian-like stroma, high columnar mucoproducer epithelium with foveolar gastric differentiation, and dysplastic changes [2].

The identification of serous cystic tumors requires cuboidal or simple squamous epithelium and fibrous or hyalinized stroma. The typical serous cystic tumor is formed by many tiny cysts and has a honeycomb appearance [4].

Recently, a different cystic pancreatic tumor has been described that typically has a non-dysplastic cuboidal epithelium, ovarian-like stroma, and contains clear or serous fluid without mucin. Despite the presence of ovarian-like stroma, this pancreatic cyst differs from MCN because it does not have any epithelial dysplastic changes, and contains non-mucinous pancreaticobiliary cystic epithelium with serous fluid. A pancreaticobiliary phenotype is also found in intraductal papillary mucinous neoplasms of the pancreas (IPMNs) and cystic neoplasms of the extrahepatic bile duct [5].

We present the case of this newly described cystic neoplasm characterized by ovarian-like stroma and a serous-like epithelium and fluid content.

2. Case Report

A 23-year-old Hispanic woman with no significant past family and medical history sought medical attention for epigastric and left upper quadrant intermittent abdominal pain, which was present during the past 18 months. Physical examination was unremarkable. Laboratory work including complete blood count, glucose, creatinine, blood urea nitrogen, liver function tests, and lipase were normal. Abdominal contrast-enhanced computed tomography and endoscopic ultrasound showed a unilocular cystic neoplasm measuring 4.9×5.8×5.7 cm dependent of the tail of the pancreas; a cyst wall measuring 3 mm; and no communication between the cyst and main pancreatic duct. Cyst fluid analysis reported amylase 45 733 IU/L, carcinoembryonic antigen 116 ng/mL and carbohydrate antigen 19-93 271 U/mL. (Figure 1).

MCN was suspected and laparoscopic distal pancreatectomy and splenectomy were performed with margin between tumor and cut surface of 2 cm. (Figure 2). Pathological examination showed a cystic neoplasm with non-mucin-producing cuboidal epithelium and ovarian-like stroma. Final diagnosis was a nonmucinous cystadenoma with pancreaticobiliary phenotype and ovarian-like stroma with negative surgical margins (Figure 3).

The patient was discharged 4 days after surgery without complications. At a 6 month follow-up visit, she was asymptomatic.



Figure 1. Evaluation of the patient by computed tomography. Axial view showed a unilocular cystic neoplasm dependent of the tail of the pancreas

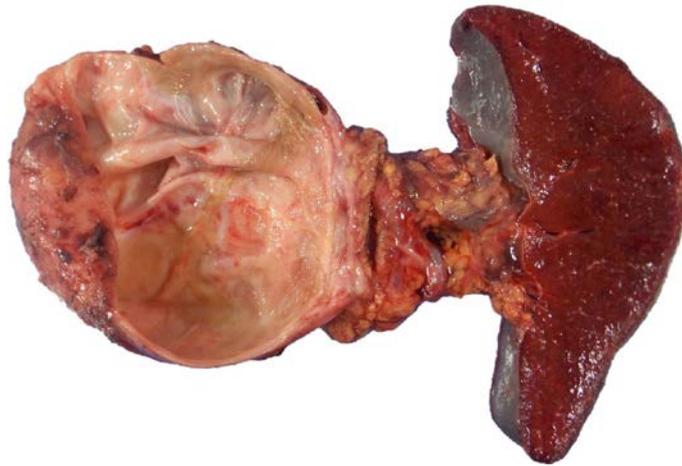


Figure 2. Surgical specimen of laparoscopic distal pancreatectomy and splenectomy. Cyst is observed in the pancreatic tail

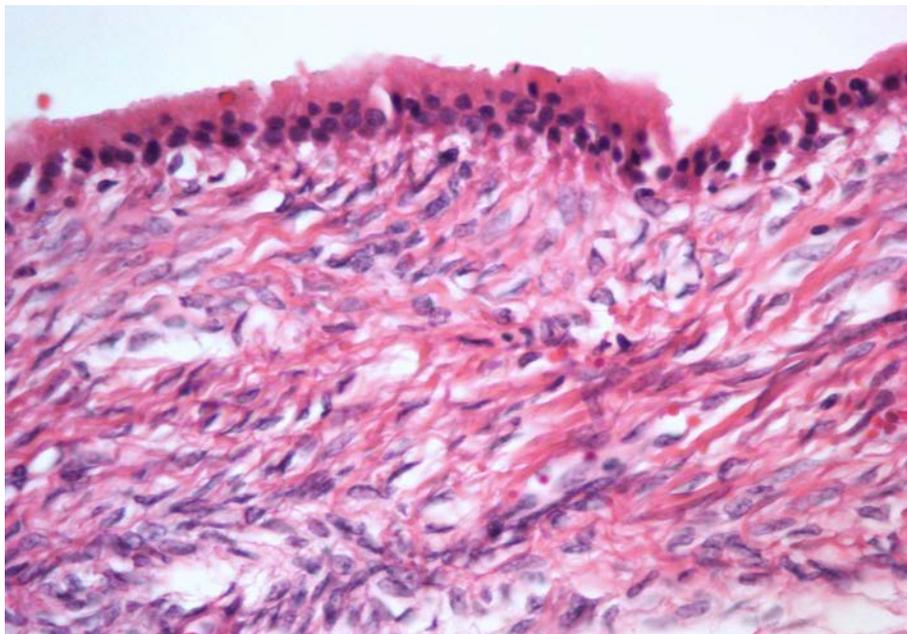


Figure 3. Nonmucinous cystadenoma of the pancreas with pancreatobiliary phenotype. Cuboidal epithelium and ovarian-like stroma are shown (Hematoxylin-eosin stained slide)

3. Discussion

According to current international consensus guidelines for the management of IPMN and MCN of the pancreas [6], we decided upon surgical management because the cyst was >3 cm, and cyst-related symptoms were present and MCN was suspected. Pathological examination revealed the presence of serous cystadenoma-like cyst

epithelium and content surrounded by MCN-associated ovarian-like stroma. This latter cystic neoplasm is not considered in the WHO classification and it has been recently described in a series of nine cases from two institutions. In that series, the neoplasms predominated in young women with a mean age of 45 years (range, 24-64 years) and their size ranged from 1.7 to 7 cm (average 4.8cm) [5] (Table 1).

Table 1. Previous published cases

Case	Sex	Symptoms	Localization	Nondysplastic cuboidal epithelium	Ovarian-like stroma
1	Female	Symptomatic	Tail and body	Present	Present
2	Female	Symptomatic	Tail	Present	Present
3	Female	Symptomatic	Tail	Present	Present
4	Female	Symptomatic	Tail	Present	Present
5	Female	Symptomatic	Tail	Present	Present
6	Female	Symptomatic	Tail	Present	Present
7	Female	Symptomatic	Tail	Present	Present
8	Female	Incidental findings	Tail	Present	Present
9	Female	Incidental findings	Tail	Present	Present

These patients are similar to our case, long-term outcome and prognosis is uncertain owing to the short-term follow-up reported in our patient. However, the lack of dysplastic changes and the absence of literature reports regarding cystadenocarcinoma arising in this specific cystic neoplasm allowed us to consider potential benign behavior. Nevertheless, we cannot overlook the possibility of early incidental diagnosis before malignant changes were present. Information from longer follow-up is needed.

To date, the presence of ovarian-like stroma has been considered synonymous with MCN. However, the present and previous reports prove otherwise, highlighting the need for a thorough examination of any pancreatic cyst before making any therapeutic decisions. Clinical manifestations, imaging characteristics, and cyst fluid analysis (with all its limitations) should be considered as part of the diagnostic approach to any pancreatic cystic lesion. It is a matter of debate whether these 10 cases are sufficient to call for a revision and the introduction of this new cystic neoplasm into the WHO classification.

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